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CREUTZFELDT-JAKOB DISEASE and BOVINE SPONGIFORM ENCEPHALOPATHY IN IRELAND.



(1980 -2000)

June 1, 2000
S. Molloy, M.D.
St. Vincent's Hospital

BSE in Ireland.

- 1989.
- BSE made notifiable disease (imported meat bone meal from UK banned).
- **1990:** all meat bone meal banned
- controls on UK imports (prohibited)
- 1996 slaughtered all birth cohorts, progeny and UK imports.
- Total 474 cases in 11 years, (mostly indigenous - 15 imports).

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Control Measures

- If animal suspected of BSE
 - herd are quarantined
 - BSE cannot be ruled out
 - suspect slaughtered
 - brain examined
 - BSE confirmed
 - entire herd slaughtered in special non-export meat factory
 - progeny traced and slaughtered
 - birth cohorts traced and slaughtered



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Specified Risk Material (SRM)

- Excluded from human and animal food chains
 - spleen of sheep and goats
 - skull, brain, eyes, tonsils, spinal cord of cattle, sheep and goats over 12 months
 - Daily audits by Dept. of Agriculture on dedicated SRM rendering plants.
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UK and Portugal

- entire head excluding tongue, thymus, spleen, intestines from duodenum to rectum and cord if >6 months
- vertebral column (including DRG) of animals >30 months in UK; >6 months in Portugal.



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Control

- 1. Ante mortem examination
- 2. Removal of SRM
- 3. Inactivation of any possible BSE agent in SRM free offal
- 4. Prevention of cross contamination of ruminant feedstuffs with SRM



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BSE(1996-2000)

• 1995	16
• 1996	73
• 1997	77
• 1998	79
• 1999	95
Jan-March	27
• 2000	
Jan-March	35



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North & South

- Border counties have higher incidence of BSE
- 1989-97:

	No. cases	Incidence
Border	69	0.84
Rest	196	0.31

- Border counties = Donegal, Monaghan, Cavan, Louth and Leitrim



Totals (1989-97)

- Republic: 265
- Northern Ireland: 1,766
- Great Britain: 170,885
- Different farming practices -
 - grass for milk and calf production
 - lower dependency on ruminant-derived protein feeds in calf rearing
 - imported ruminant ration from North (mainly used in border counties)

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1996 European Collaboration.

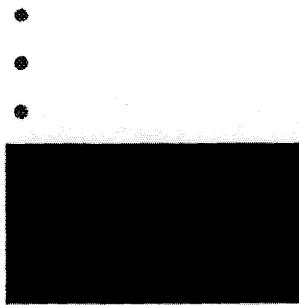
- **Objectives:**
- clinicopathological classification criteria
- co-ordinate surveillance programmes
- analyse risk factors for vCJD
- harmonise methods of data collection
- accurate epidemiological information

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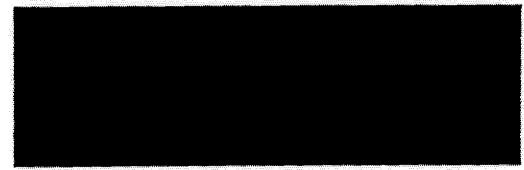
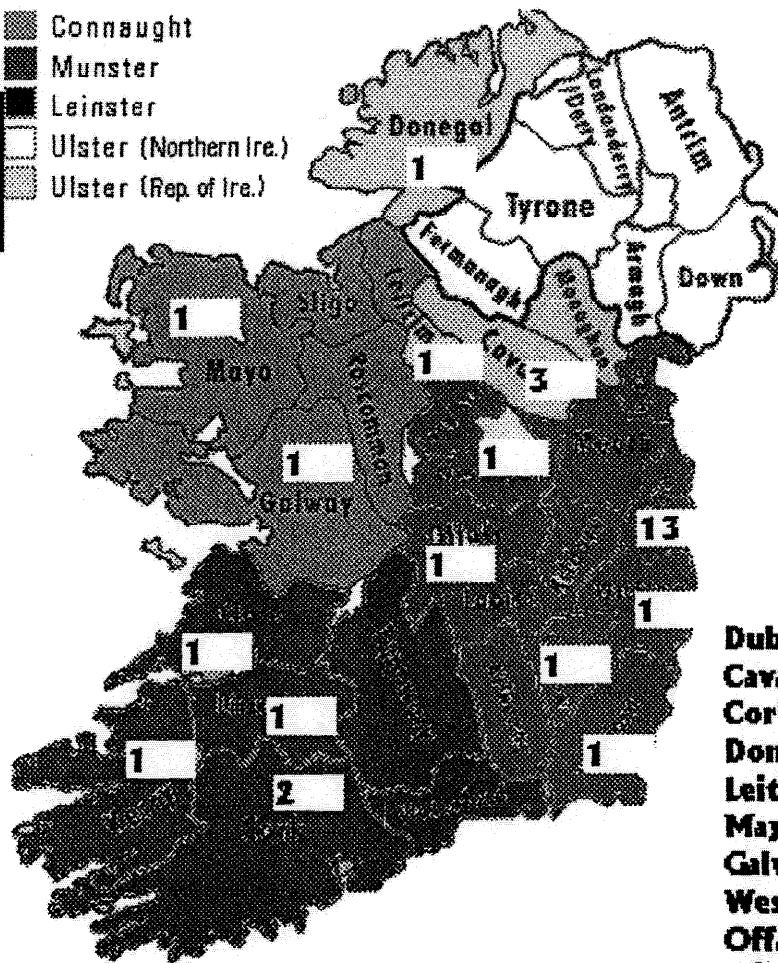
IRELAND(1980-96)

- **Retrospective Study**
- death certs
- neuropathological data from Cork and Dublin.
- 20 cases identified (13 women, 7 men)
- data on 15.
- Annual mortality of 0.31 per million





Connacht
Munster
Leinster
Ulster (Northern Ire.)
Ulster (Rep. of Ire.)



Dublin	13
Cavan	3
Cork	2
Donegal	1
Leitrim	1
Mayo	1
Galway	1
Westmeath	1
Offaly	1
Wicklow	1
Wexford	1
Carlow	1
Limerick	1
Clare	1
Kerry	1

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1996

- **notifiable disease December 1996.**
 - active surveillance; suspected cases referred notified to Neuropathology Lab in Cork
 - CJD Advisory Committee
 - Dept.'s of Health and Food & Agriculture
 - Monthly meetings
 - Post mortems done in two centres - Cork and Beaumont
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CJD data since May 1996

- 12 cases definite CJD (neuropathological confirmation).
- data on 11.
- equal sex incidence .
- 1 genetic mutation at codon 178 causing fatal insomnia ?familial,



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Features

- **Age of onset:** 58 (32-78)
- **Duration of illness:** 21.5 weeks
- **Presentation:** 100% dementia
- **Risk factors?:**
 - 2/11 significant time in the UK,
 - 1 meat processor,
 - 1 leather factory worker.



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Active Surveillance

- July 1999; formation of active surveillance unit with
 - clinical referral centre in St. Vincent's Hospital, Dublin,
 - neuropathological referral centre in Beaumont Hospital.
- Notification of all physicians, neurosurgeons and psychiatrists of the same.



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Active Surveillance (July 1999...)

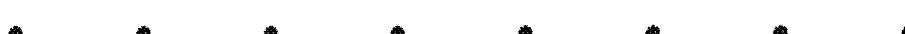
- 10 referrals; 4 clinical, 6 neuropathological
- 5 women, 5 men
- 7 RIP - 2 sporadic CJD
 - 1 gliomatosis cerebri
 - 1 FTD
 - 2 pending PM
- 1 VARIANT



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Variant case report

- 33 year old mother of two
- painful right leg
- unsteady
- depressed
- Collateral (husband):
 - confirmed depression
 - confusion (delusions & hallucinations)
 - vivid dreams
 - deteriorating memory



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- Past history: Nil
- Family history: Adopted
- Social History: Married, 2 children,
Chef,
Midlands of Ireland
UK resident for 6 years
(1989 - 1995)
- Blood donor in the UK



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Examination

- Cognition; poor recall (1/4)
WORLD
serial sevens (93)
- Cerebellum; mild limb ataxia
severe truncal ataxia

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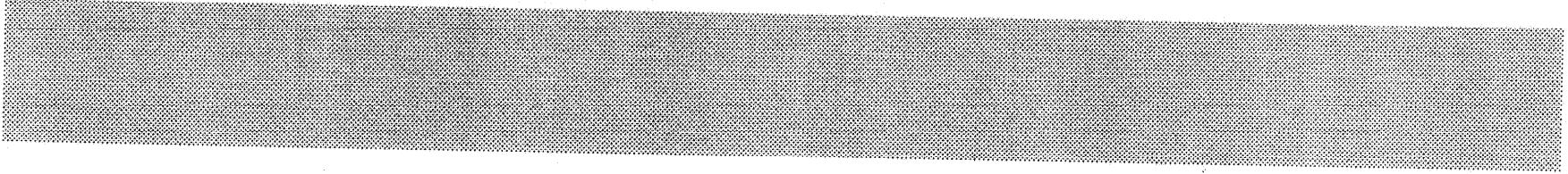
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Investigations

- Haem & Biochem: normal
- CT brain: normal
- CSF: protein & glucose normal
C&S/ AFB/ viral: nil
immunoglobulins; normal
14-3-3 positive
- EEG: non specific abnormalities



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- **MRI brain;** high signal abnormalities in posterior and medial aspects of the thalamus bilaterally.
 - Codon 129 : M/M
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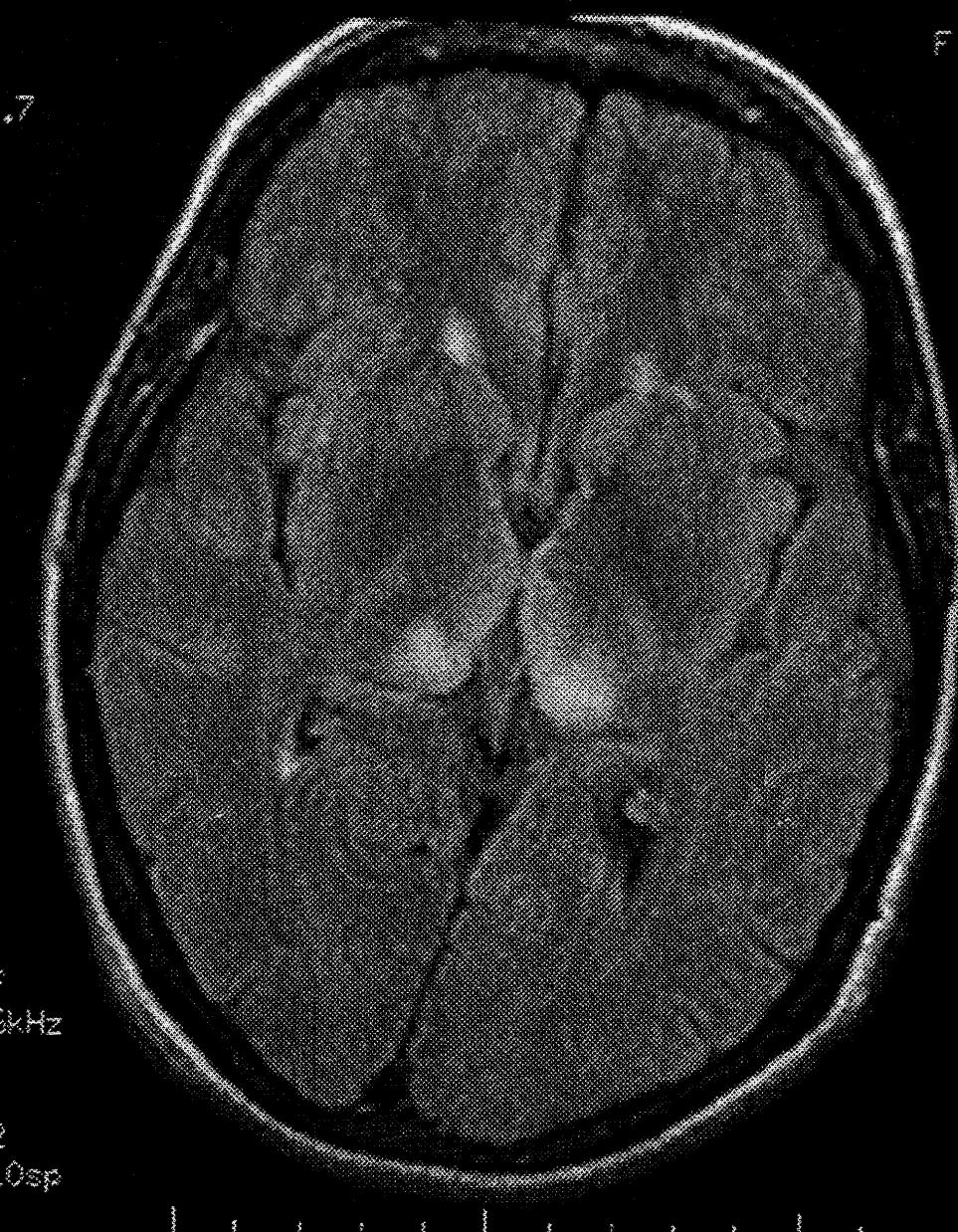
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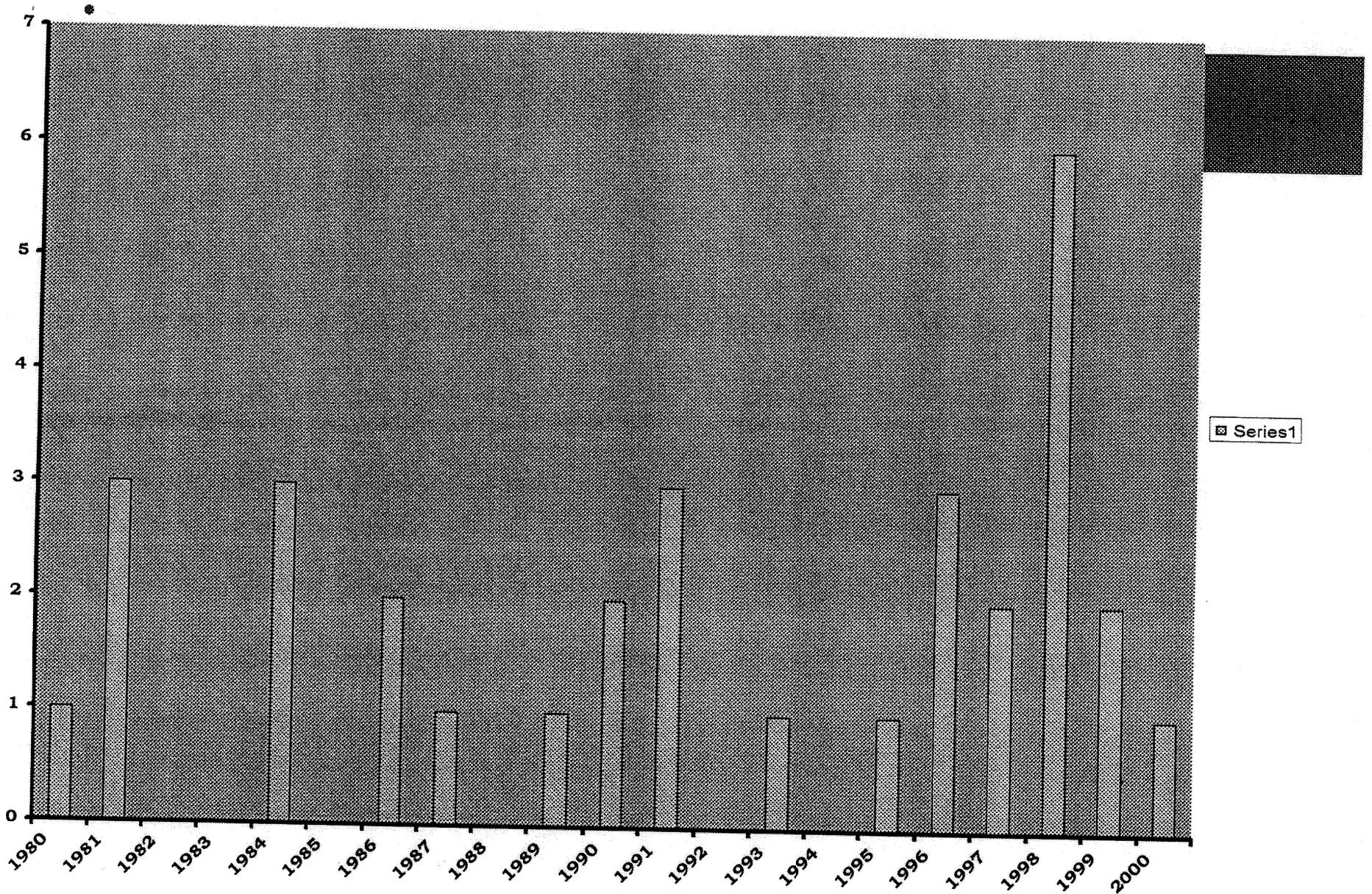
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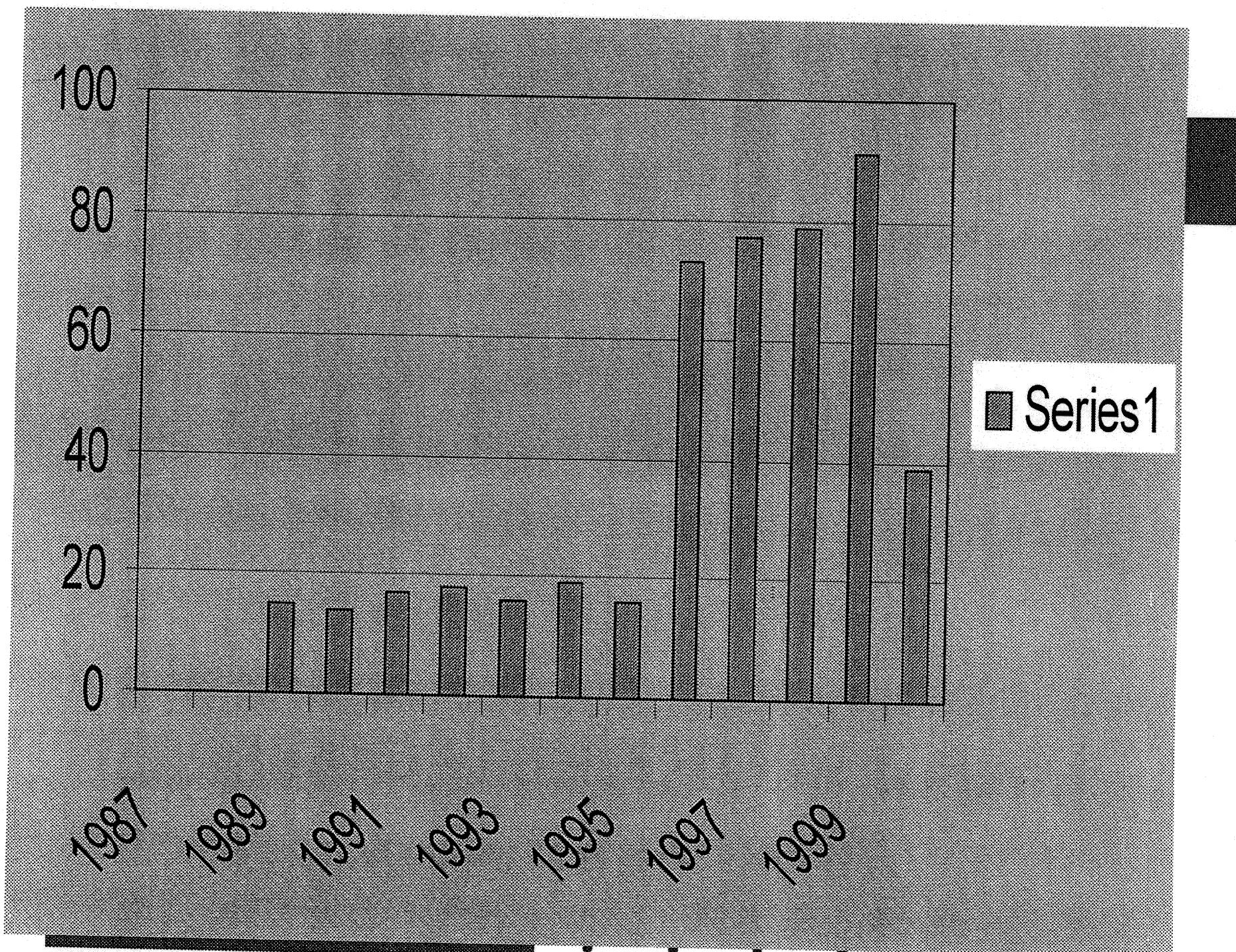
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Combined Data (CJD in Ireland between 1980-2000).

- 32 cases/20 years/pop. 3.8×10^6 (19/32 definite)
- annual mortality 0.44/yr/m
- **UNDER ASCERTAINMENT??**



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Problems with Surveillance of CJD

- Uncertainties/ variations in case definition.
- Developing diagnostic techniques
- Variable diagnostic policies
- Factors triggering notification
- Geographical variation in risk factors
- Links to policy makers



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Problems in Ireland

- Rare disease
 - Public awareness
 - Dementia in the elderly
 - Agricultural based economy
 - Diagnosis by post mortem
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